CLAIMS

1. A process for the preparation of a composition or a protein suitable for maintaining intact and/or restoring and/or increasing the number of cellular mitochondria, i.e., for modifying in a positive sense the activity of production of intracellular energy, characterized in that it envisages the use, as active ingredients, of the branched amino acid leucine, or a pharmaceutically acceptable derivative thereof, in combination with at least one between the branched amino acids isoleucine and valine, or pharmaceutically acceptable derivative thereof.

2. The process according to Claim 1, in which leucine, isoleucine, and valine, or pharmaceutically acceptable derivatives thereof, are used as active ingredients.

3. The process according to Claim 1 or Claim 2, in which the ratio between the amount of isoleucine and the amount of leucine, or derivatives thereof, on molecular weight basis, is from 0.2 to 0.7, preferably from 0.4 to 0.6.

4. The process according to Claim 1 or Claim 2, in which the ratio between the amount of valine and the amount of leucine, or derivatives thereof, on molecular weight basis, is from 0.2 to 0.7, preferably from 0.4 to 0.6.

5. The process according to at least one of the preceding claims, in which at least one between threonine and lysine, or a pharmaceutically acceptable derivative thereof, is envisaged as further active ingredient.

6. The process according to Claim 5, in which the ratio between the amount of threonine and the amount of leucine, or derivatives thereof, on molecular weight basis, is from 0.15 to 0.50, preferably from 0.2 to 0.45.

7. The process according to Claim 5, in which the ratio between the amount of lysine and the amount of leucine, or derivatives thereof, on molecular weight basis, is from 0.15 to 0.60, preferably from 0.3 to 0.55.

8. The process according to Claim 5, in which threonine and lysine, or derivatives thereof, are both present as further active ingredients and the sum of their amounts is between 10% and 50%, preferably between 25% and 45%, of the sum of the amount of leucine, isoleucine and valine, or derivatives thereof, on molecular weight basis.

9. The process according to one or more of the preceding claims, in which there are envisaged, as further active ingredients, one or more essential amino acids, or pharmaceutically acceptable derivatives thereof, selected in the group

consisting of histidine, methionine, phenylalanine, tryptophan.

10. The process according to Claim 9, in which there are envisaged, as further active ingredients, histidine, methionine, phenylalanine, tryptophan, or pharmaceutically acceptable derivatives thereof.

11. The process according to Claim 10, in which the sum of the amounts of histidine, methionine, phenylalanine, tryptophan, or derivatives thereof, is from 2% to 25%, preferably from 5% to 15%, of the sum of the amount of leucine, isoleucine, valine, threonine and lysine, or derivatives thereof, on molecular weight basis.

12. The process according to Claim 9, in which at least one between tyrosine and cyst(e)ine, or a pharmaceutically acceptable derivative thereof, is provided as further active ingredient.

13. The process according to Claims 9 and 12, in which tyrosine, or derivative thereof, is in an amount from 15% to 50%, preferably from 20% to 35%, of the amount of phenylalanine, or derivative thereof, on molecular weight basis.

14. The process, according to Claims 9 and 12, in which the amount of cyst(e)ine, or derivative thereof, is at least 100%, and preferably comprised between 150% and 350%, of the amount of methionine, or derivative thereof, on molecular weight basis.

15. The process according to Claim 12, in which one or more further amino acids, or pharmacologically acceptable derivatives thereof, are envisaged as active ingredients, the sum in gram molecular weight of which is in a percentage lower than 20% with respect to the other active ingredients and/or less than 10% for each individual further amino acid, or derivative thereof.

16. The process according to at least one of the preceding claims, in which the sum of the individual amounts of threonine and lysine, or derivatives thereof, on molecular weight basis, is smaller than the sum of the individual amounts of the aforesaid branched amino acids, or derivatives thereof, but greater than the sum of the individual amounts of the other essential amino acids, or derivatives thereof, envisaged in the composition.

17. The process according to at least one of the preceding claims, in which the amount of threonine, or derivative thereof, on molecular weight basis, is smaller than the individual amounts of lysine and of the aforesaid branched amino acids, or derivatives thereof, but greater than the individual amounts of the other

essential amino acids, , or derivatives thereof, envisaged in the composition.

18. The process according to at least one of the preceding claims, in which the amount of lysine, or derivative thereof, on molecular weight basis, is smaller than individual amounts of the aforesaid branched amino acids, or derivatives thereof, but greater than the individual amounts of the other essential amino acids, or derivatives thereof, envisaged in the composition.

19. The process according to at least one of the preceding claims, in which the amount of threonine, or derivative thereof, on molecular weight basis, is smaller than the individual amounts of lysine and of the aforesaid branched amino acids, or derivatives thereof, but greater than the sum of the individual amounts of the other essential amino acids, or derivative thereof, envisaged in the composition.

20. The process according to at least one of the preceding claims, in which the amount of lysine, or derivative thereof, on molecular weight basis, is smaller than the individual amounts of the aforesaid branched amino acids, or derivatives thereof, but greater than the sum of the individual amounts of the other essential amino acids, or derivatives thereof, envisaged in the composition.

21. The process according to at least one of the preceding claims, in which the composition has a pH in an aqueous solution comprised between 6.5 and 8.5, with or without excipients suitable for the preparation of tablets, capsules, powders, etc., in any pharmacological form of presentation suitable for enteral or parenteral use.

22. Use of a composition or a protein obtained with the process according to one or more of Claims 1 to 21 for the production of a formulation for the treatment of pathological conditions distinguished by insufficient or reduced mitochondrial function, such as:

- degenerative diseases of the nervous system, and in particular Alzheimer's disease, amyotrophic lateral sclerosis, Parkinson's disease;
- apoptosis of mitochondrial origin;
- ischaemic states that inhibit the energetic activity of the cell;
- sarcopenia of the aged;
- senescence.

23. Use of a composition or protein obtained with the process according to one or more of Claims 1 to 21 for the production of a formulation suitable for increasing neuronal activity in degenerative illnesses of the nervous system.

24. Use of a composition or protein obtained with the process according to one or more of Claims 1 to 21 for the production of a formulation suitable for the maintenance of the structures designed to produce cellular energy, and in particular for the maintenance of the integrity of the mitochondria.

25. Use of a composition or protein obtained with the process according to one or more of Claims 1 to 21 for the production of a formulation suitable for use in any condition where an improved peripheral use of oxygen is advantageous in patients with progressive loss of cells consequent to apoptosis mediated by the mitochondria, in particular via activation of caspase-9.

26. Use of a composition or protein obtained with the process according to one or more of Claims 1 to 21 for the production of a formulation suitable for antagonizing the apoptotic phenomena of the mitochondria and controlled by the mitochondria, activated by caspase-9.

27. Use of a composition or protein obtained with the process according to one or more of Claims 1 to 21 for the production of a formulation suitable for bringing about an increase in the energetic activity of cells, and hence of the tissue and organ that the set of cells composes, such as an increase of the neuronal activity in the degenerative diseases of the nervous system, in which the activity of the cells decreases on account of reduced energetic capacity that involves a loss of the mitochondrial energetic activity.

28. A protein obtained via genetic engineering or other artificial method, having a composition of amino acids according to one or more of Claims 1 to 21.

29. Use of at least one of the branched amino acid leucine, isoleucine and valine, or a pharmaceutically derivative thereof, as active ingredient for the preparation of a composition or a protein suitable for maintaining intact and/or restoring and/or increasing the number of cellular mitochondria, i.e., for modifying in a positive sense the activity of production of intracellular energy.

30. Use of at least one of the branched amino acid leucine, isoleucine and valine, or a pharmaceutically derivative thereof, as active ingredient for the production of a composition or a protein for the treatment of pathological conditions distinguished by insufficient or reduced mitochondrial function, such as:

- degenerative diseases of the nervous system, and in particular Alzheimer's disease, amyotrophic lateral sclerosis, Parkinson's disease;
- apoptosis of mitochondrial origin;

- ischaemic states that inhibit the energetic activity of the cell;
- sarcopenia of the aged;
- senescence.

31. Use according to claim 29 or 30, wherein at least one of threonine and lysine, or a pharmaceutically derivative thereof, is provided as further active ingredients of the composition or protein.